

Diagnosis of Epilepsy

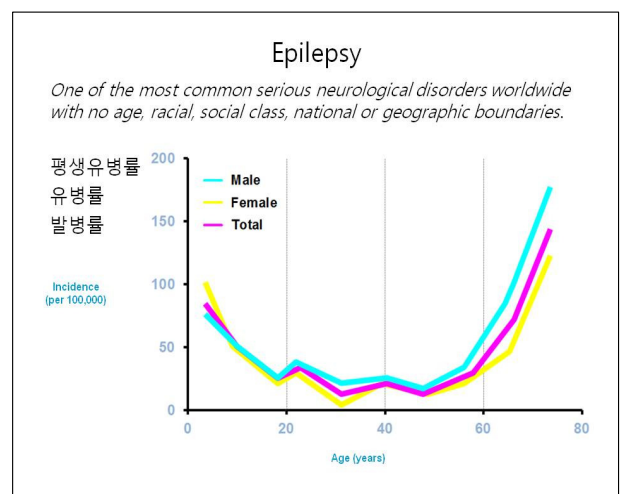


서 대 원

성균관대학교 의과대학 신경과



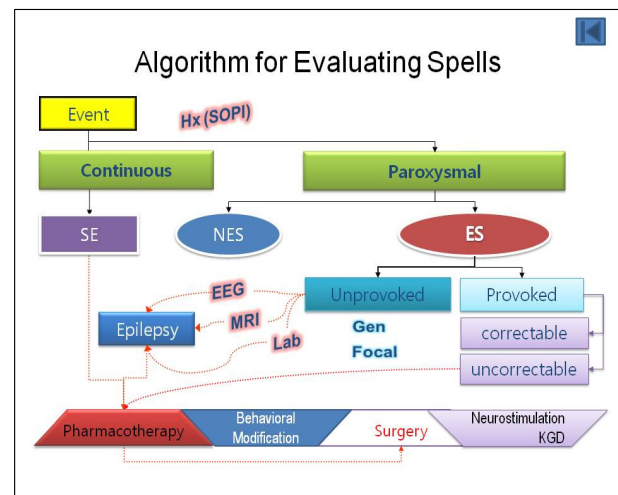
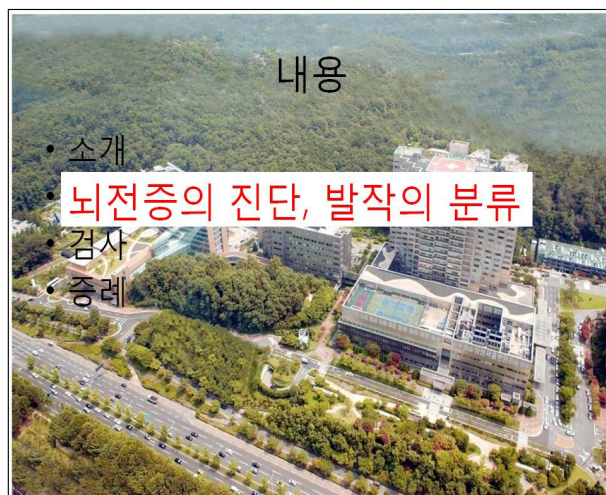
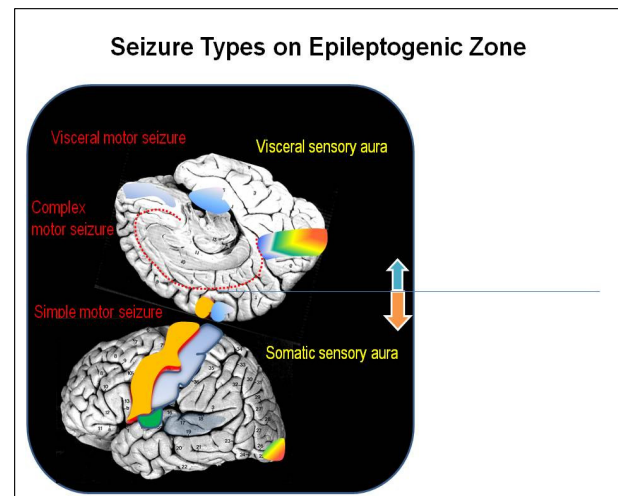
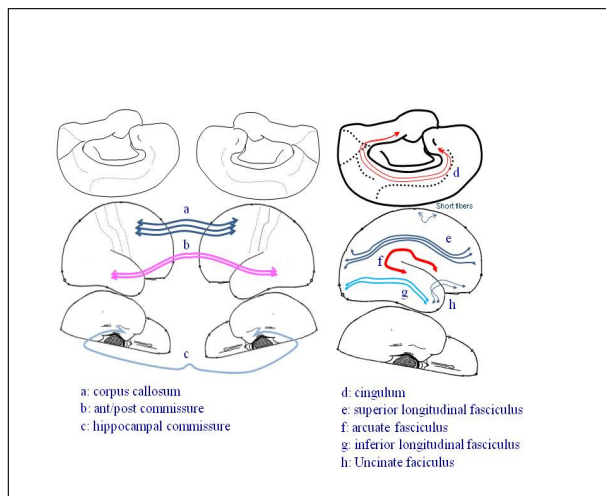
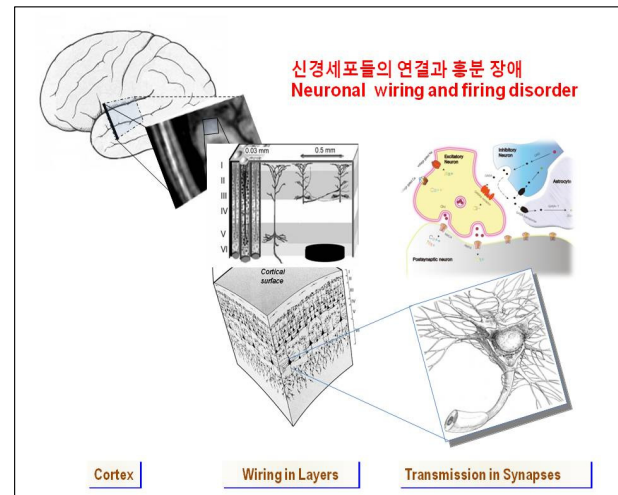
용어			
우리말	한자	일본어	영어
경련	痙攣	けいれん(게이렌)	Convulsion
발작	発作	ほっさ(홋샤)	Seizure
간질	癇疾		Epilepsy
전간	癲癇	てんかん(텐칸)	



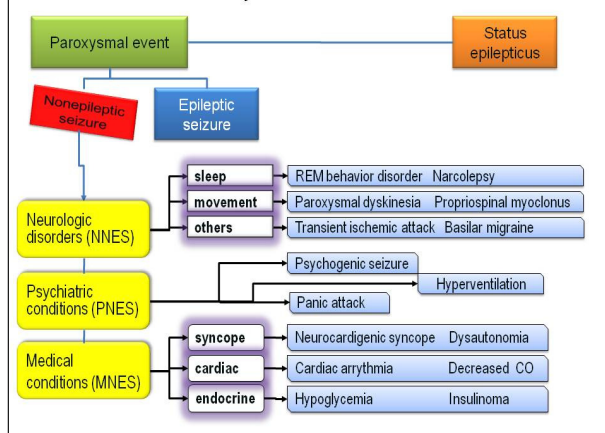
Definition

- **Epilepsy** is
a common chronic neurological **disorder**
characterized by recurrent unprovoked **seizures**.^{[1][2]}
- The epileptic seizures are
transient signs and/or symptoms of
abnormal, excessive or synchronous neuronal activity in the brain.^[3]
- The brain disorder is characterized by
an enduring predisposition to generate epileptic seizures and by
neurologic, cognitive, psychological, social consequences of this condition.^[3]

1. Commission on Epidemiology and Prognosis, ILAE. Epilepsia 1989.
2. Blume W, et al. Glossary of descriptive terminology for ictal semiology report of the ILAE task force on classification and terminology. Epilepsia 2001.
3. Fisher R, et al. Epileptic seizures and epilepsy: definitions proposed by the ILAE and IBE. Epilepsia 2005.



Classification of Paroxysmal Events



Epilepsia, 51(4):671-675, 2010
doi: 10.1111/j.1528-1167.2009.02285.x

SPECIAL REPORT

Recommendation for a definition of acute symptomatic seizure

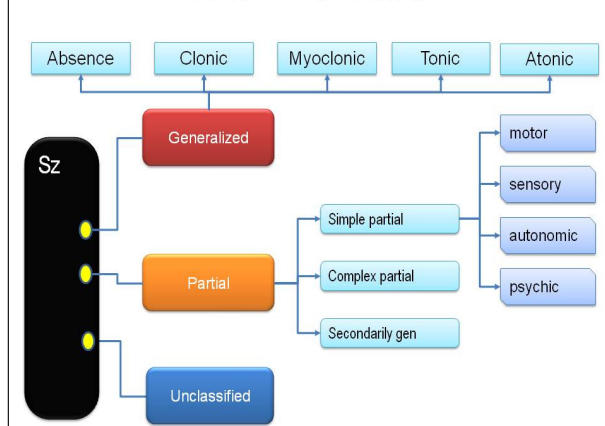
*Ettore Beghi, †Arturo Carpio, ‡Lars Forsgren, §Dale C. Hesdorffer, ¶Kristina Malmgren, #Josemir W. Sander, **Torbjorn Tomson, and ††W. Allen Hauser

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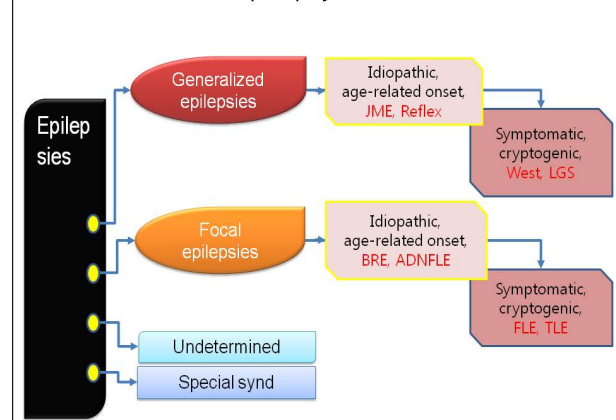
within the first 7 days of Specific Causes

CVD	Biochem. parameter	Value
TBI, including intracranial	Serum glucose	<36 mg/dl or
CNS infections		>450 mg/dl w ketoacidosis
Cerebral hypoxia	Serum sodium	<115 mg/dl
within 7–48 h of	Serum calcium	<5.0 mg/dl
the last drink	Serum magnesium	<0.8 mg/dl
within 24hr of	Urea nitrogen	<100 mg/dl
metabolic toxic systemic causes	Creatinine	>10.0 mg/dl

발작(seizure) 국제분류



간질(Epilepsy) 국제분류



Epilepsia, 51(4):676-685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

SPECIAL REPORT

Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005–2009

*†Anne T. Berg, ‡Samuel F. Berkovic, §Martin J. Brodie, ¶Jeffrey Buchhalter, #**J. Helen Cross, ††Walter van Emde Boas, ‡‡Jerome Engel, §§Jacqueline French, ¶¶Tracy A. Glauser, ##Gary W. Mathern, ***Solomon L. Moshé, †††Douglas Nordli, †††Perrine Plouin, and ††Ingrid E. Scheffer

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Table 1. Classification of seizures^a

Generalized sz

Tonic-clonic (in any combination)

Absence

Typical

Atypical

Absence with special features

Without impairment of consciousness or awareness

With observable motor or autonomic components

This roughly corresponds to the concept of "simple partial seizures."

"Focal motor" and "autonomic" are terms that may convey this concept depending on the seizure manifestations.

Involving subjective sensory or psychic phenomena only

This corresponds to the concept of an *aura*, a term endorsed in the 2001 Glossary.

With impairment of consciousness or awareness

This roughly corresponds to concept of complex partial sz.

"Dyscognitive" is a term that has been proposed for this concept (Blume et al., 2001).

Focal sz

Unknown Evolving to a bilateral convulsive^b seizure

(involving tonic, clonic, or tonic and clonic components).

This expression replaces the term "secondarily generalized seizure."

Table 3. Electroclinical syndromes and other epilepsies

Electroclinical syndromes arranged by age at onset*

Neonatal period
 Infancy
 Childhood
 Adolescence – Adult

Juvenile absence epilepsy (JAE)
 Juvenile myoclonic epilepsy (JME)
 Epilepsy with generalized tonic-clonic seizures alone
 Progressive myoclonus epilepsies (PME)
 Autosomal dominant epilepsy with auditory features (ADEAF)
 Other familial temporal lobe epilepsies

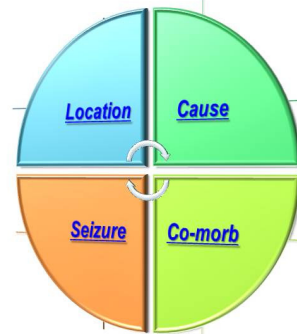
Less specific age relationship
 Familial focal epilepsy with variable foci (childhood to adult)
 Reflex epilepsies

Distinctive constellations
 Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
 Rasmussen syndrome
 Gelastic seizures with hypothalamic hamartoma
 Hemiconvulsion-hemiplegia-epilepsy

Epilepsies attributed to and organized by structural-metabolic causes
 Malformations of cortical development (hemimegalencephaly, heterotopias, etc.)
 Neurocutaneous syndromes (tuberous sclerosis complex, Sturge-Weber, etc.)
 Tumor Infection Trauma Angioma Perinatal insults Stroke etc

Epilepsies of unknown cause
 Benign neonatal seizures (BNS), Febrile seizures (FS)

Four Dimensions of Epilepsy Classification



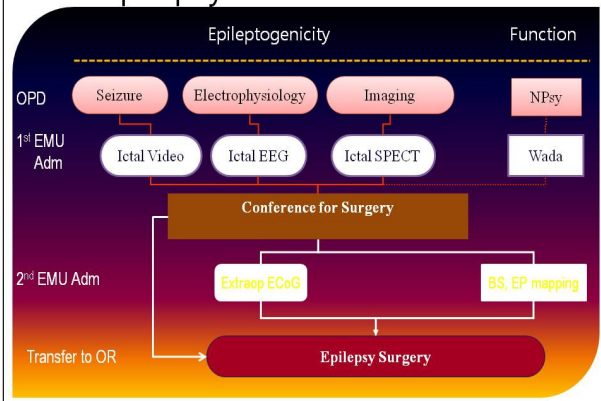
Example of Classification

Epileptogenic *left temporal lobe (mesial temporal)*
Causes *neoplasm (left mesial temporal DNT)*
Seizures
 semiology *abdominal aura → automotor seizure*
 frequency *persistent (1 seizure/m)*
 triggering *none*
Conditions *severe verbal memory problems*

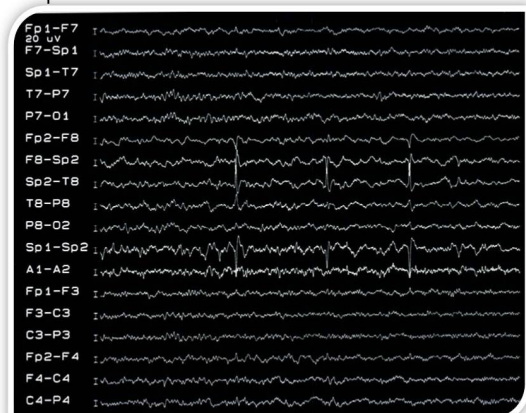
내용

- 소개
- 진단
- 뇌전증의 진단적 검사
- 증례

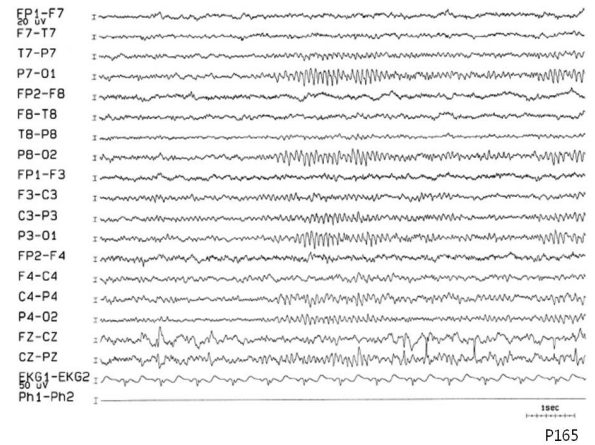
Epilepsy Patient Evaluation



Sphenoidal Maximum INIC ED in MTLE



C/IED in FCD



EEG

Typical Epileptiform Discharges

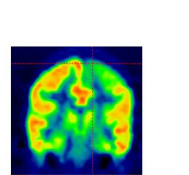
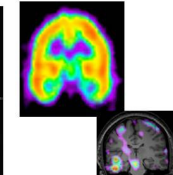
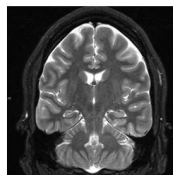
Epilepsy	EEG pattern (interictal)	Epileptic syndrome (Etiology)
Focal	Anterior temporal spike	MTLE
	Reg. Psp (C/IED)	Focal cortical dysplasia
Generalized	Gen. 3Hz SWC	Absence epilepsy
	Gen. >4Hz PSpWC	JME
	Hypsarrhythmia	West syndrome
	Gen. slow SWC	LGS

Neuroimaging

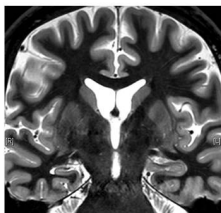
MRI

SPECT

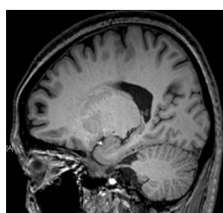
PET



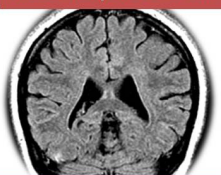
MRI



Mesial Temporal Sclerosis



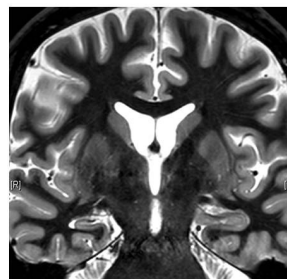
Malformation of Cortical Development



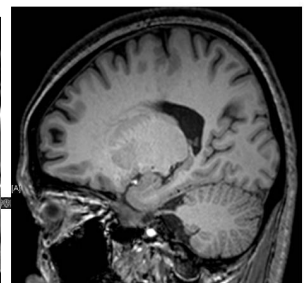
Encephalomalacia



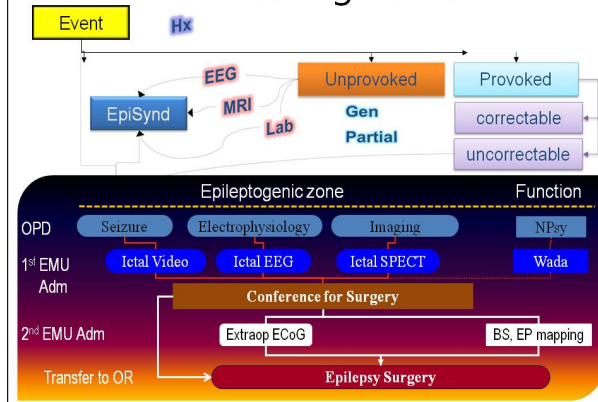
Neoplasm



Mesial Temporal Sclerosis



Malformation of Cortical Development





Case 1. 김H신 44/F/ 18ya

87년도, 18yo (고3, 87년도)

갑자기 멍해지면서 의식을 잃고 온몸에 힘이 빠지면서 휘청 간혹 넘어지기도 하며,

주로 왼팔/손에 힘이 빠지면서 잡고 있던 것을 놓침. 1/wk

95년도, 출산 후 2일 전신발작. 1년 지나 산후 우울증 입원 중 1회 더 당시 왼팔에 힘 빠지는 느낌이 들면서 왼팔이 흔들리고

전신발작으로 넘어가는 양상. AED복용시작

07년도, 본원 외래 f/u 왼팔 힘 빠짐 1/wk, 전신발작 1/y

14년도, EMU monitoring

.	OXC	LTG	LEV	VPA
07.10.31	150-300	100-100		
08.03.24	"	150-150		
08.09.03	150-450	"		
09.02.16	"	162.5-162.5		
11.05.27	150-375	"		
12.02.03	225-450	"		
12.09.04	"	100-150		
13.01.28	"	150-150		
13.05.31	"	"	500-500	
13.06.12	"	"	1000-750	
13.06.28	"	"	1500-750	300
13.07.12	"	"	"	300-150
13.09.27	225-150-300	25-150	"	"

Diagnosis

Focal Cryptogenic Epilepsy

Cause : unknown

Seizure

Semiology : atstatic seizure

Lt. arm ss aura → Lt. arm clonic → GTC

Frequency : special seizure 1/wk

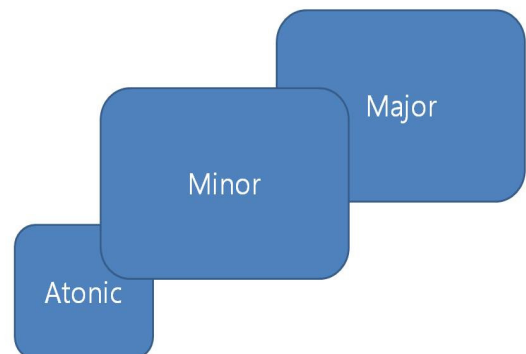
SGS total 2

Triggering : fatigue, emotional stress

Comorbidity : migraine without aura

Video Clip

Psychogenic Nonepileptic Seizures



문1. Nonepileptic psychogenic seizure의 type으로 tremulous movement가 포함되는 유형은?

- 가) major motor
- 나) minor motor
- 다) negative motor

Case 2. 이H현민 29/F 19ya

2008(중3) 앉아 공부 중 화장실 가려고 일어나는데,
갑자기 왼팔 → 왼다리 저리며, 뒤통지이며 뻗뻗느낌
10초. 그후 갑자기 소실.
앉았다 일어나면서 발생, 드물게 주저앉기까지 함.
버스, 지하철, 도서관에서 자주 발생, 집 드림. 1-3/d

2009.02.14 PED 내원 EMU monitoring.
INIC ED: no IC ED: theta to delta in both FC

2009~2011 AED 복용에 뚜렷한 호전 없음
이후 self D/C AED. 증상은 지속

2014.01.10 NR 내원. OXC 300 M. 이후 2-3회/일 → 1회/일
2014.01.22 EMU 재입원

EMU at Ped F/15 10yo

1. Clinical History
가. Seizure onset: 08.3월경
나. Semiology : Type I) 왼쪽 팔이 떨리고 다리에 힘이 빠진다.
dur: 10-20sec post ictal Sx.: none frequency: 1-2/dat

다. Past medical history : none
라. AED before epilepsy monitoring unit admission: none

2. Video EEG monitoring Result
가. Days of monitoring : 2 days
나. Description and number (2회)
(1) Sz #1.2: numbness of Lt extremity
→clonic sz of Lt arm (sensory aura→Lt side motor seizure)
EEG: theta to delta in both frontocentral area

다. 상세기술 (including lateralizing signs) (1) Lt side clonic seizure
라. Interictal spikes total = none
마. Features of ictal EEG (1) Lateralized to none (2) Regionalized to FC

3. PLAN oxcarb 300mg bid opd f/u

PED NEURO 000/000

EMU at NR F/20 10yo

Monitoring 20140122~20140124 Electrodes: Scalp FT 10-10

1.Video EEG monitoring Result
-Interictal abnl: none
-Clinical events (5회)
(1) Description -S01~05: 자리에서 일어난 17초~33초 후
→왼팔(다릴) 저린느낌→힘빠짐 →왼손꼬임
(2) Ictal EEG -S01-05: no EEG change

2. Diagnosis: r/o partial seizure << r/o Paroxysmal nonkinesigenic dysknesia

3. Plan
AEDs: 입원 전 퇴원시
Oxcarbazepine 300mg(M)
Carbamazepine D/C 300mg(M)

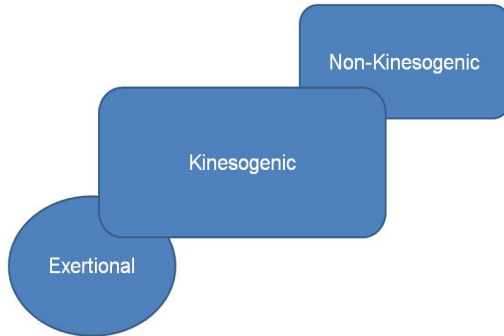
황경진/서대원

AED History

AED	OXC	LTG	LEV
09.02.27	600-600		
09.03.20	450-450		
09.05.28	600-600		
09.06.18	"	37.5- 37.5	
09.07.31	"	87.5- 87.5	
09.10.30	"	100 - 100	
10.01.08	"	137.5- 137.5	
10.02.19	"	162.5- 162.5	
10.04.23	"	187.5- 187.5	
10.05.24	450-450	200 - 200	
10.06.25	300-300	"	
10.07.23	"		500 - 500
10.10.07	"		1000 -1000
11.04.08	"		" - 마지막 PED 처방일
14.01.10	300 M		

Video Clip

Paroxysmal Dyskinesias



문2. Paroxysmal dyskinesia로 epileptic cause로 보이는 것은?

- 가) Kinesogenic choreoathetosis
- 나) Non-kinesogenic dyskinesia
- 다) Exertional induced dyskinesia
- 라) Hypnic dyskinesia

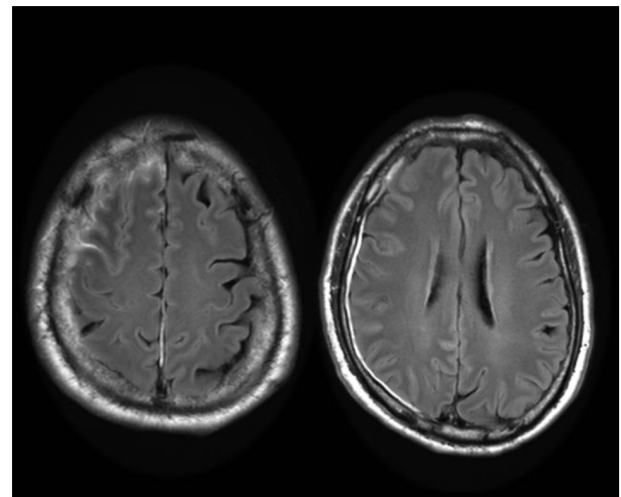
Case 3. 박M환 M/59

2013.12.12
NR Ha 진료 중 발작.
눈앞 흔들거리는 어지럼→고개좌측→운동뻘뻘. 1분. 서서히 갸.
ER EEG: WNL.
MRI: R.frontal cortical nodular lesion r/o glioma f/u MRI 3m

2013.12.20
NR Ep ■ 진단 Symptomatic focal epilepsy
FTL in rt. SFG r/o tumor
Aura → Lt. versive seizure → GS: SGS once
Rt. hemispheric ha, r/o ha ass. with medical illness

■ 계획 NS 협진, Levetiracetam 500mg bid 35일

NR Ha ■ 계획 [투약] ◆1 Propranolol 40mg 0.5T 2P 35일
◆1 Flunarizine 5mg 1C 2P 35일
[검사] C-spine AP/lat/ob, TCD, Routine【비급여】

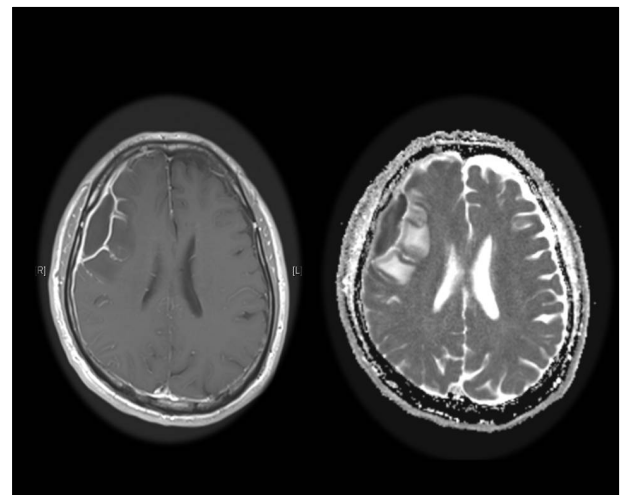


2013.12.24 NS
■ 진단 Benign neoplasm of brain, supratentorial,
Acute gastric ulcer without haemorrhage or perforation

■ 계획 [투약] Utracet(R)ER 650mg/75mg 1T 2P 30일
Afloqualone 20mg 1T 2P 30일
Rabeprazole 10mg 1T M 30일

[검사] 3달후 MRI (PRE- & POST-CONT) + Diff + Perf (비급여)
→ 2013.12 진단시 급여 적용. 다음 급여 2014/12, 15, 17, 19 급여 적용후 종료

2013.12.30 ER
두통에 대해 NR, NS OPD f/u 하며 약 복용중. 호전 없는 상태로 지냄.
내원 1일전 일상생활 도중 nausea, vomiting 발생. 두통 이전과 동일.
전일 점심때부터 금일 오전까지 vomiting 4차례, nausea 지속되어 ER.
화장실갈때 Lt. side tilting tendency 있어 ER visit
Rt.Frontal convex 의 fluid collection 에 대하여 NS contact



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SPECIAL REPORT

Recommendation for a definition of acute symptomatic seizure

*Ettore Beghi, †Arturo Carpio, ‡Lars Forsgren, §Dale C. Hesdorffer, ¶Kristina Malmgren, #Josemir W. Sander, **Torbjorn Tomson, and §W. Allen Hauser

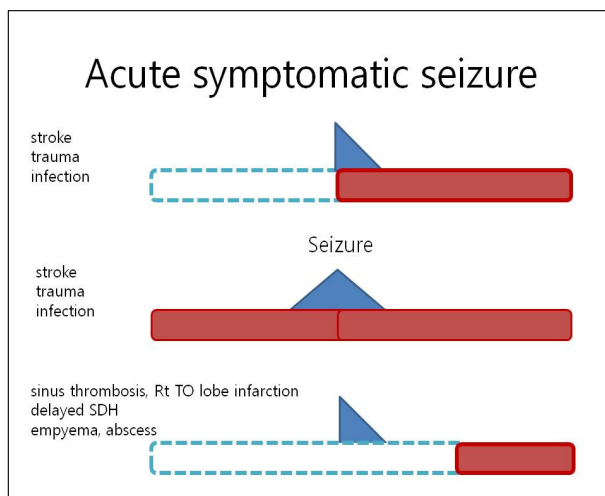
*Mario Negri Institute, Milan, Italy; †Department of Neurology, University of Cuneo, Cuneo, Ecuador; ‡Department of Neurology, Umea University, Umea, Sweden; §Department of Epidemiology and Department of Neurology, Columbia University, New York, New York, U.S.A.; ¶Göteborg University, Göteborg, Sweden; #UCL Institute of Neurology, Queen Square, London, United Kingdom and SEIN - Epilepsy Institute of the Netherlands Foundation, Heemskede, The Netherlands; and **Karolinska Institutet, Stockholm, Sweden

within the first 7 days of
Specific Causes
CVD;
TBI, including intracranial;
CNS infections.
Cerebral hypoxia

Operatively
within 7~48 h of the last drink
within 24hr metabolic toxic systemic causes

Proposed cutoff values for acute symptomatic seizures in common metabolic disorders

Biochemical parameter	Value
Serum glucose	<36 mg/dl (2.0 mM) or >450 mg/dl (25 mM) ass. with ketoacidosis (whether or not there is long-standing diabetes)
Serum sodium	<115 mg/dl (<5 mM)
Serum calcium	<5.0 mg/dl (<1.2 mM)
Serum magnesium	<0.8 mg/dl (<0.3 mM)
Urea nitrogen	<100 mg/dl (>35.7 mM)
Creatinine	>10.0 mg/dl (>884 IM)



문3. Acute symptomatic seizure인 것은?

가) 발작 직후 serum-Na 121 mg/dl
나) ACA infarction 후 14일째 발작
다) meningitis 후 8일째 발작
라) 금주 후 36시간째 발작

Case 4. 이|Y| 32/F 5YA

1. History

2007.10.18 캐나다 토론토 어학연수 중 김치 볶음밥해 프라이팬 들고가다 갑자기 멎어지며 정면응시하고 불려도 대답없고 혼자서 말하는 모습. 혼동에서 회복후 왼손이 아파서 보니까 화상입음

2007.11. 한강성심병원 GS 왼손 화상 피부이식위해 입원.
MRI: L pons, R MB, B cbl, L thal, L GP, R hippo, R amygd lesions
Szs: 갑자기 정면 주시하다 찢쩍. 오른손 만지작.

2008.04.01 본원 NS 입원. 뇌병변 r/o multifocal glioma로 navigation-guided biopsy. Path: no tumor cells.
그후 CPS (1~2/월) 발생. AED polytherapy로도 발작 지속됨.

2012.08.07 본원 NR 거쳐 presurgical evaluation 위해서 EMU 입원.

2. Semiology

Abd or vertig aura → automotor seizure, Rt hemi; SPS 1-2/w, CPS 1-2/m

3. AED before EMU admission

	CBZ	TPM	LEV	PGB	CBZM
2008.08.12	500-500				
2012.07.17	500-500	200-200	1500-1500	75-75	5-5

Video-EEG monitoring results (20120807~20120809)

INIC ED: Sp, 1-2/2-3min, T8(100.0%)

IC events (11회)

nausea(2) → lip smacking(1,7,9,6,8) → R EB dev(3,10,11) → L fumbling (4,5)
Ictal EEG Rt temporal onset (10/11), No EEG change (1/11)

Neuroimaging

B-MRI (2012-08-07): R hippocampomegaly Multifocal infiltrative lesions
SPECT(2012-08-09; #11, 40/102sec: injection시 R EB dev; B temp rhythm): Hyperperfusion in the right temporal cortex

PET (2012/10/10): Hypometabolism in R DL frontal and ant to mid temp

Epilepsy classification: Right temporal lobe epilepsy, symptomatic

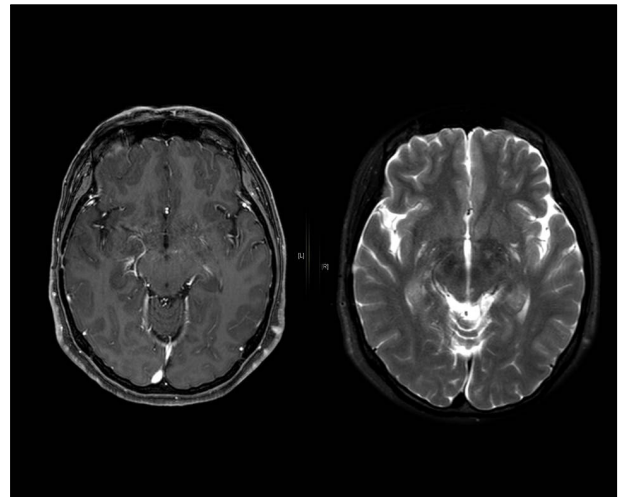
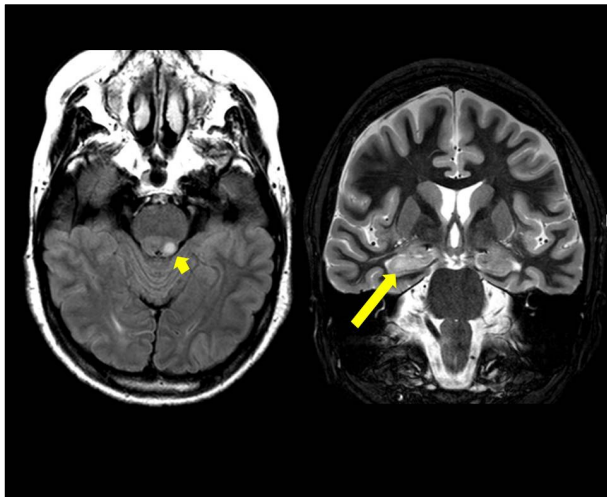
Seizures: abdominal aura → automotor seizure

Cause: Rt hippo. lesion R/O CNS lymphoma, R/O NMO, R/O vasculitis

Related medical conditions: none

Plan CNS lymphoma, NMO 등의 감별필요
right TLE에 대한 수술적 치료도 고려함.
AED는 CBZ 500mg bid → OXC 750mg bid로 교체함.

전임의 김지영/ 교수 서대원



NF1
café-au-lait spots, inguinal/axillary freckling, neurofibromas.

- Diagnosed by two of the following C/F
 - ≥ 6 café-au-lait spots (>1.5 cm in adults)
 - ≥ 2 neurofibromas of any type or plexiform
 - Freckling under the arms or in the groin
 - ≥ 2 Lisch nodules (iris hamartomas)
 - optic nerve tumor (glioma)
 - osseous lesion (sphenoid dysplasia)
 - relative with NF1 by the above criteria
- Additional but not diagnostic features:
 - Precocious puberty or delayed sexual developm
 - learning disabilities in reading, spelling or math
 - Growth
 - Macrocephaly
 - Scoliosis
 - Hypertension
 - Epilepsy

TSC
Adenoma sebaceum, shagreen patch, hypomelanotic macules

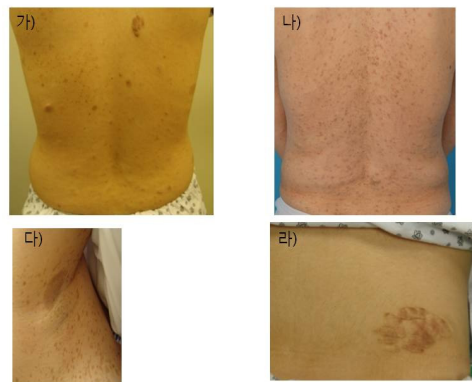
Major Features

- Facial angiofibromas or forehead plaque
- Non-traumatic unguinal or periungual fibroma
- Hypomelanotic macules (≥ 3)
- Shagreen patch (connective tissue nevus)
- Multiple retinal nodular hamartomas
- Cortical tuber, Subependymal nodule, Subependymal giant cell astrocytoma
- Cardiac rhabdomyoma, single or multiple
- Lymphangiomyomatosis
- Renal angiomyolipoma

Minor Features

- "Confetti" skin lesions
- Retinal achromic patch, Multiple randomly distributed pits in dental enamel, Gingival fibromas, Cerebral white matter migration lines
- Hamartomatous rectal polyps, Bone cysts, Non-renal hamartoma, Multiple renal cysts

문 4. TSC를 의심할 수 있는 피부병변은?



Summary

